INDIAN PACING AND ELECTROPHYSIOLOGY JOURNAL 15 (2015) 296–299

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Bidirectional ventricular tachycardia of unusual etiology

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ARTICLE INFO

Article history:
Available online 23 February 2016

ABSTRACT

Bidirectional ventricular tachycardia (BDVT) is a rare form of ventricular arrhythmia, characterized by changing QRS axis of 180 degrees. Digitalis toxicity is considered as commonest cause of BDVT; other causes include aconite toxicity, myocarditis, myocardial infarction, metastatic cardiac tumour and cardiac channelopathies. We describe a case of BDVT in a patient with Anderson-Tawil syndrome.

Case report

An 11 years old boy was brought with complaints of exercise induced palpitation. There was no associated dyspnoea, chest discomfort or syncope. He was hemodynamically stable. Physical examination was unremarkable except for tachycardia. ECG (Fig. 2) showed bidirectional ventricular tachycardia with right bundle branch block morphology. Tachycardia was terminated with combination of oral beta blocker and Flecainide. ECG in normal sinus rhythm (Fig. 3) showed normal QT interval with prominent U wave in V3–V4. There was no family history of similar complaints or history of sudden death in family. There was no history of substance abuse or drug intake. There was no obvious dysmorphic features except small proximal phalanges of little fingers of both hands, giving an impression of fifth finger clinodactyly (Fig. 1). The child's height was normal as per his age and mid-parental height. There was no history suggestive of muscle weakness. Routine biochemical investigations including serum electrolytes, troponin T and BNP were normal. Echocardiography revealed normal bi-ventricular function and structurally normal heart. Differential diagnosis of Catecholaminergic Polymorphic Ventricular Tachycardia and Anderson-Tawil syndrome (ATS) were entertained. The patient was sent for neuro-electrophysiological examination. Routine nerve conduction studies and electromyography were normal. Exercise testing, carried out after prolonged exercise, showed initial increase in compound muscle action potential (CMAP) amplitude with a progressive drop and slow recovery. In view of genetic heterogeneity for bidirectional VT, genomic DNA was subjected to exome sequencing by Next Generation

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Peer review under responsibility of Indian Heart Rhythm Society.
http://dx.doi.org/10.1016/j.ipej.2016.02.007

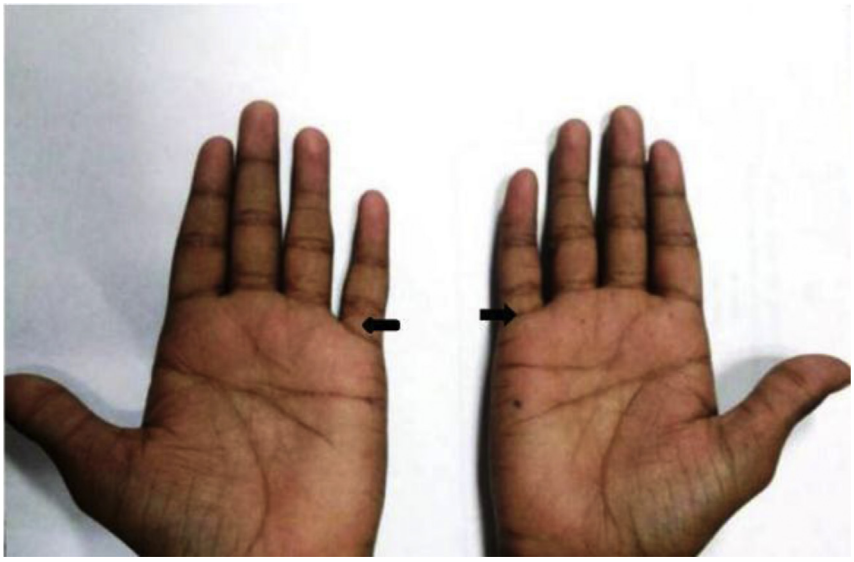

Fig. 1 – Hands showing small proximal phalanges of both little fingers (Arrow).

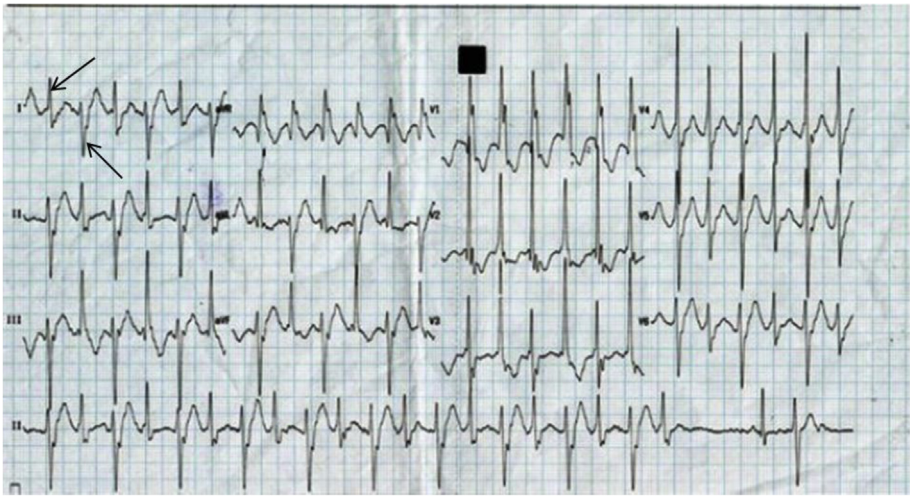

Fig. 2 – ECG showing Bi-directional Ventricular Tachycardia with RBBB morphology and alternate change in frontal axis (Arrow).

sequencing (NGS) technique. Genes responsible for catecholaminergic polymorphic ventricular tachycardia (CPVT) and Anderson-Tawil syndrome were analysed for sequence variations. The patient was found to harbour a known, pathogenic, heterozygous variant p.Arg218Trp caused by a substitution (c.652C > T) in exon 2 of the KCNJ2 gene. Both parents were screened for the same variation by Sanger sequencing. None of them was found to harbour this variation, suggesting that the variation is denovo in the affected child.

Discussion

A variety of clinical conditions have been implicated in the generation of bidirectional VT. Apart from digitalis toxicity, other etiologies include myocarditis, myocardial infarction [1], metastatic cardiac tumour [2], herbal aconite poisoning and cardiac channelopathies i.e. catecholaminergic polymorphic VT (CPVT) and Anderson-Tawil syndrome (ATS). Absence of drug or herbal medicine intake, structurally normal heart with normal bi-ventricular function pointed towards Channelopathy in our patient. In ATS, unlike CPVT, ventricular arrhythmias are associated with extra cardiac manifestations like episodic flaccid muscle weakness and dysmorphic features [3]. Characteristic dysmorphologies include low-set ears, hypertelorism, small mandible, clinodactyly and syndactyly. Our patient did not have any of the described dysmorphic features except fifth finger clinodactyly. In 60% patients (also called Type 1 ATS) pathogenic mutation in KCNJ2 gene is evident. Reduced IK1 resulting from KCNJ2 mutations alters late cardiac repolarization and leads to both distinctive T-U wave morphology and an increased propensity to ventricular arrhythmias. In a series by Haruna et al. [4], 30% patients

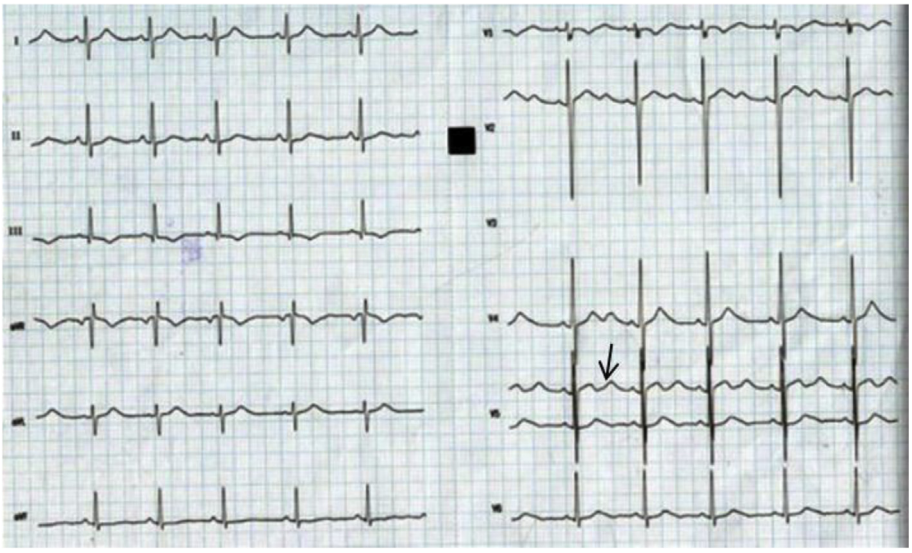

Fig. 3 – ECG in Sinus Rhythm showing prominent U wave (Arrow).

presented with classical triad of symptoms and 70% patients presented with any two of three symptoms. Characteristic T-U–wave pattern is present in 90% of Type 1 ATS and this pattern has 84% sensitivity and 97% specificity in diagnosing ATS [5]. Ventricular tachyarrhythmia has been observed in about 65% patients, bidirectional VT being most common followed by polymorphic VT, monomorphic VT and rarely VF [4]. Cardiac conduction abnormalities [5] and dilated cardiomyopathy [6] has also been reported.

In absence of flaccid muscle paralysis, long exercise protocol may reveal an immediate post -exercise increment followed by an abnormal decrement in the compound motor action potential after prolong exercise [7], as in our patient.

Differentiation of ATS from CPVT is crucial because later situation is much more fatal than former and sudden cardiac death is rare in ATS. Although ATS can be differentiated from CPVT by classical extra-cardiac features, there is a probability of phenotypic expression of cardiac derived symptomatology only, as in our case. However a nonfatal course, prominent U wave with prolongs QU interval and subclinical neuroelectrophysiological finding pointed towards ATS.

ATS is inherited in an autosomal dominant manner. At least 50% patients diagnosed with ATS have an affected parent. Up to 50% of cases present sporadically without family history and these cases are due to denovo mutation, as in our case [8]. The mutation also carries high degree of variable phenotypic expression and incomplete penetrance, with as much as 20% mutation positive carriers being asymptomatic [9].

Despite high arrhythmia burden, patients with KCNJ2 Mutation (ATS I) have low rate of syncope and cardiac arrest. No single pharmacological agent has been found to be effective in preventing arrhythmia load. While few patients showed improvement with calcium channel blockers or Amiodarone, same drugs are ineffective in other cases [10] or even exacerbate torsades de pointes [11]. Although beta-blockers and sodium channel blockers (mexiletine, propafenone, and flecainide) have been described ineffective [10], a good response was seen in our patients with combination of beta-blocker and flecainide. Pacing in combination with nicorandil has been described to be effective in some cases [11]. Apart from reducing the arrhythmia load by pharmacotherapy, implantable cardioverter-defibrillator (ICD) is indicated to prevent sudden cardiac death (SCD). Although presence of ATS itself is considered a class IIb indication for ICD placement, history of cardiac arrest due to ventricular fibrillation or VT should be regarded as a class I indication for ICD placement in ATS [11]. Until further data regarding risk stratification becomes available, ICD therapy should be reserved for ATS patients with a history of cardiac arrest, syncope, or sustained rapid and/or symptomatic VT [11].

To conclude we report a case of BDVT of rare etiology and diagnosis of ATS should be considered in presence of BDVT with structurally normal heart, even in absence of classical diagnostic criteria.

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